

## **Giant frontal and cerebellopontine angle epidermoid cyst. Case report**

**M. Dabija, N. Ianovici**

Neurosurgery, University of Medicine and Pharmacy “Gr.T. Popa” Iasi

### **Abstract**

We present the case of a 38-years old woman presenting in a state of unconsciousness, (Glasgow Coma Scale score 7), with right hemiparesis, the onset of symptoms being acute, after 3 consecutive convulsive crisis, in a patient with no previous neurological sign, except progressive aggravating headaches. Cranial computed tomography reveal an enormous hypodense lesion in the left frontal and temporal lobe, frontal horns of the ventricular system and prepontine and cerebellopontine angle regions. The patient underwent emergent surgical intervention, the evacuation of the content and partial removal of the capsule of the giant epidermoid cyst was achieved by frontal craniotomy. The diagnosis was presumed by the classical aspect of the content of the cyst and it was confirmed by histological examination. The immediate postoperative evolution was good with regression of hemiparesis and comatose state, with one complication in the third day after surgery; she developed the symptoms of Mollaret meningitis, with nearly normal CSF parameters, who responded well at the treatment with dexamethasone.

**Keywords:** epidermoid cyst, aseptic meningitis

### **Introduction**

Epidermoid cysts, also known as "pearly tumors" because of their bright white appearance at intraoperative resection, developed from epithelial inclusions in basal cisterns, diploe of the skull and, very rarely in the intrinsic brainstem or pineal region, during neural tube closure or formation of secondary cerebral vesicles. They are not really tumors, their growth is due to the continuous formation of the keratohyalin from continued desquamation by the epithelium tissue. Their growth is very slow, so the onset of their clinical manifestations is variable between 20 and 40 years in most of the cases.

We present our experience with a patient who has a giant epidermoid cyst in

the frontal area with infratentorial extension in the prepontine and cerebellopontine angles with surgical resection of the lesion. We report these very rare case and we made a short discussion with reviews of the literature.

### **Case report**

We present our experience with the case of a 38 years old woman who presented in the emergency room in a comatose state, with right hemiparesis, right facial central nerve paresis, generalized hypertonic state with neck stiffness and bilateral Babinsky sign. It was in fact a post epileptic condition, the patient suffered 5 consecutive crisis with no free interval between them. The

mother of the patient reported a history of chronic headaches with a progressively aggravated evolution, which was, especially in the last two month, intractable to non-narcotic analgesics, associated occasionally with nausea, dizziness and vomiting.

A CT-scan was performed in emergency and it revealed an extremely large well circumscribed hypodense lesion, which occupied the anterior left cranial fossa, the temporal pole, involved the both frontal horns with infratentorial extension in the prepontine and cerebellopontine cisterns. The formation was slightly hyperdense when compare to the CSF and it had a heterogeneous appearance. No enhancement was noticed after contrast administration.



Figure 1 Preoperative image of the frontal-temporal cyst



Figure 2 The lesion involves the frontal horns of the lateral ventricles



Figure 3 Prepontine extension of the epidermoid cyst



**Figure 4** Right cerebellopontine extension of the lesion

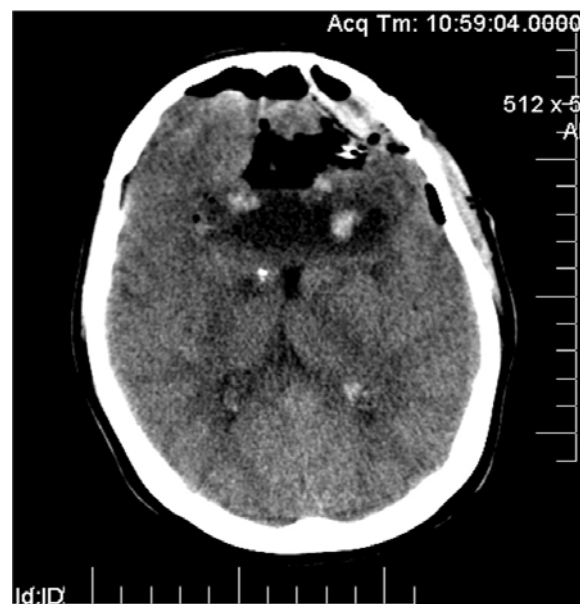
The patient was hospitalized in the intensive care unit and the evolution was favorable with the recovery of the state of conscience but with a mild confusion and somnolence and persistence of the right hemiparesis and dysphasia.

The very next day the patient underwent the surgical intervention. We performed a fronto-temporal craniotomy and, after the incision of the dura we discovered the specific aspect of the pearly-white desquamated keratin flakes which was removed and the dissection of the capsule was performed. This capsule was very adherent especially at the level of the sylvian and carotid vessels and at the level of the optic nerve and even at the cortical level in some points, therefore we was forced to make only a subtotal excision. We demanded a histological exam of the capsule and that one confirmed a stratified squamous epithelium. The postoperative CT\_scan

revealed a remnant of the epidermoid material at the level of the ventricular system, especially in the right (contra lateral) one. Because of the very low growing rate of the lesion we decided to survey these remnants and those of the posterior fossa.



**Figure 5** Postoperative image at 24 hours



**Figure 6** Postoperative image at 48 hours



**Figure 7**  
**Postoperative**  
**image:**  
**ventricular**  
**residual**  
**epidermoid**  
**material**

The postoperative evolution was very good in the first two days with complete recovery of the preoperative mild hemiparesis, but, in the third day, the patient accused severe headaches, nausea, vomiting, and fever (temperature between 38 and 39,5°C). We performed a lumbar puncture and that one revealed no meningitis aspects, WBC count of 0/mm<sup>3</sup>, glucose was 50mg/dl, the proteins was 76 mg/dl and we found 2560 red cells/mm<sup>3</sup>. The aseptic Mollaret meningitis was very probable the cause and the evolution was good with the remission of the symptoms in three days under the treatment with dexamethasone. In the fourth and fifth postoperative days the patient installed two right jacksonian crisis and we start a treatment with Depakine chrono 1500mg/day. The patient was discharged with no neurological deficits and no other accuses in the 14-th postoperative day.

## Discussions

Epidermoid tumor was first reported in 1683 by Duverney(7) and their first epidermal origin description and their nomination as “epidermoid” was in 1854 by Remak(8). Epidermoid is a term who means “epiderm like” and it clearly

suggests that their content is made of the tissues normally present in the skin. The capsule of these lesions is, in fact, a keratinizing stratified squamous epithelium that resembles the keratinizing epidermis of the skin and the fibrous capsule is a dermal connective tissue (1). The interior is formed by a “pearly” white, waxy tissue, formed by concentric lamellar deposits of keratin rich in cholesterol crystals, secreted by the basal cuboidal germinative stratus of the peripheral epidermal tissue (2). Their origin is in ectopic inclusions of epithelial cells during neural tube closure (3) or as a dysembriogenesis of the gastrulation with a secondary disruption of the neural tube continuity in the third to fifth week of gestation (4).

The location of the epidermoid cysts includes the diploic region of the calvaria, ventricular system, cerebellopontine angle, optic chiasm and parapituitary region, pineal area and the collicular plate. They can displace or envelope neural or vascular structures and they may have a thick capsule witch can be very adherent to the surrounding brain tissue, vessels and nerves.

Due to their very slow growth, their mean age of clinical manifestations and hospital admission is 35 years and they occur more often in women (5). The dermoid cysts, witch contains also hair follicles and sebaceous glands, tend to reveal by more precocious symptoms, because their content, more heterogeneous, increase more rapidly.

One of the most common initial symptoms is the severe headache due to the raised intracranial pressure but, very often, to the aseptic meningitis, explained by the leakage of the cholesterol in the

cerebrospinal fluid and chemical irritation (6). That may be the case of our patient who suffered intractable headache with nausea and vomiting in the last two months before the presentation.

The symptoms are not specific and depend on the location of the cyst. The clinical picture developed insidiously due to the slow keratin formation but there are also described acute evolutions by rupture of the cysts (spontaneous, traumatic, or iatrogenic) or by cystic hemorrhage. The rupture of the cyst can produce also the aseptic meningitis or Mollaret meningitis. Mollaret described that type of recurrent, aseptic and self-limited meningitis in 1944 in a patient with an spinal thoracic epidermoid lesion. He described also the Mollaret cells, a type of leucocytes larger than the common PMNs with round or kidney shape and deep situated nucleus, presented in the blood tests of that patient (9). Our patient installed that type of meningitis in the postoperative period, with violent headaches, nausea, vomiting, fever, neck stiffness photophobia and nearly normal constants in the CSF analysis. The symptoms responded well to corticoid therapy. Ventricular involvement, with both frontal horns occupied by the epidermoid material may play a role in the preoperative violent headaches and postoperative Mollaret meningitis.

### Conclusions

Epidermoid cyst is a rare condition, and the predilection for the ventricular system or cerebellopontine angle is well known, but we present a case which associated the two previous locations with the frontal temporal location, an

extremely rare one. The onset of symptoms with progressive aggravating headaches, nausea and vomiting is the common one, but the presentation of the patient with 5 consecutive generalized crises, with hemiparesis and mild dysphasia might be a rare debut. Postoperative evolution was good with complete recovery and absence of any symptoms at the discharge from the hospital and in the next 2 years of our neurological surveillance. The postoperative aseptic meningitis is a common complication and is due to the chemical irritation of the CSF and the subarachnoid space by the collagen rich content of the cyst.

### References

1. Robert H. Wilkins, Setti S. Rengachary: Neurosurgery, vol.1, part VI, Neuro-oncology: An Overview, Section E, Epidermoid and Dermoid Tumors, pp 655-661;
2. Fornari M, Solero C L: Surgical treatment of intracranial dermoid and epidermoid cysts in children, Childs nervous system, 6: 66-70, 1990;
3. Gormley W, Tomecek F, Qureshi N: Craniocerebral epidermoid and dermoid tumors: A review of 32 cases, Acta Neurochirurgica (Wien) 128; pp1115-1121, 1994;
4. Dias M, Walker M: The embryogenesis of complex dysraphic malformations: A disorder of gastrulation? Pediatric Neurosurgery 18: pp229-253, 1992;
5. Winn R, Dacey R: Youmans Neurological Surgery, fifth edition, Saunders, Volume 1, section II, part 6, 68 Epidermoid, Dermoid and Neurenteric Cysts, 1223-1231, 2004
6. DeSouza C, DeSouza R, Da Costa S: Cerebellopontine angle epidermoid cysts: a report on 30 cases, Journal of Neurology, Neurosurgery, and Psychiatry;52:986-990, 1989;
7. Ji Young Jeon, Jae Min Kim: Epidermoid Cyst of the Fourth Ventricle, Journal of the Korean Neurosurgery Society 38: 478-480, 2005;
8. Remak R : Ein Beitrag zur Entwicklungsgeschichte der krebshaften Geschwulste. Dtsch Arch Klin Med. 6: 170, 1854;
9. Baoqin Gao, Jun Yang: Mollaret Meningitis Associated With an Intraspinal Epidermoid Cyst, Pediatrics 120;e220-e224, 2007;